CONGENITAL BOWING OF THE ULNA AND AGGRESSIVE FIBROMATOSIS

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The association of skeletal anomalies and aggressive fibromatosis has been documented. Isolated bowing of the ulna is rare, yet its occurrence, particularly in conjunction with congenital dislocation of the radial head, has been documented. This article presents two cases of ulnar bowing in which the patients subsequently developed aggressive fibromatosis. We feel that aggressive fibromatosis may be a latent manifestation of congenital bowing of the ulna. The course of the disease appears to be of an aggressive nature, and patients who present with bowing of the ulna should be followed for the potential development of this disease. (*J Natl Med Assoc.* 1991;83:978-982.)

Key words • ulna bowing • fibromatosis

Isolated bowing of a long bone in children is uncommon. Bowing involving the ulna is most frequently described in association with congenital dislocation of the head of the radius.[1,2] Trauma also has been described as an etiologic agent.[3,4] Isolated bowing of a long bone also has been described as a sequela postradiation therapy. Further isolated bowing of the distal ulna associated with a bony dysplasia has been reported.[5]

Aggressive fibromatosis is a term used to describe a controversial, poorly defined group of fibroblastic lesions that mimic malignant tumors by local proliferation and invasion, although they probably lack the capacity to metastasize. The very nature of the lesion, which involves voluntary muscles as well as aponeurotic and fascial structures and the tendency to local persistence, makes the clinical diagnosis and treatment an enigmatic challenge to the surgeon.

A Medlars search failed to reveal a case of association of congenital bowing of the ulna and aggressive fibromatosis. This article presents two case reports with long-term follow-up in which aggressive fibromatosis developed in patients with congenital bowing of the ulna.

MATERIALS AND METHODS

Case 1

A 43-year-old black male presented in October 1979 with a 5-month history of a gradually enlarging mass in the posterior aspect of the right forearm. He had noted mild discomfort in the distribution of the dorsal sensory branch of the radial nerve for approximately 1 year. A demonstrable neurologic finding was an incomplete deficit in the dorsal sensory branch of the radial nerve. There was no history of trauma. He described bowing of the ulna since childhood.

Physical examination revealed a posterior medial bowing of the right forearm and a firm, nontender mass, 4×5 cm, in the extensor compartment. Range of motion of the elbow was full, including pronation and supination. There was no local or distant lymphadenopathy.

Radiographs of the involved extremity showed an extensive lesion of the proximal ulna with reactive bone formation and cortical thickening interspersed in areas of bone resorption (Figure 1A). Bowing of the ulna was apparent. The radial head was not dislocated. Technetium-99m polyphosphate scintigraphy demonstrated

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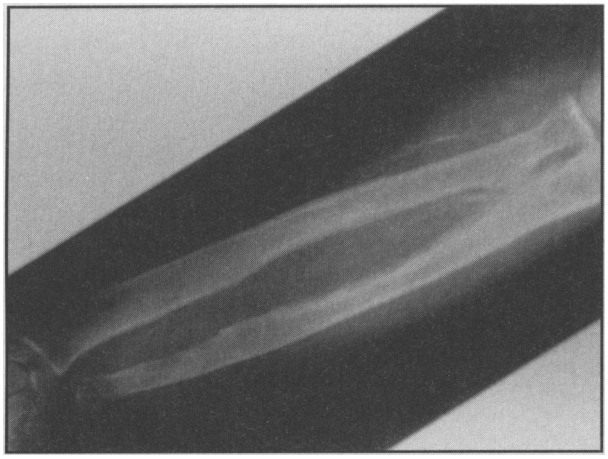

Figure 1A. Radiographs demonstrating soft tissue striations, bowing of the ulna, and marked cortical destruction of the ulna diaphysis (Case 1).

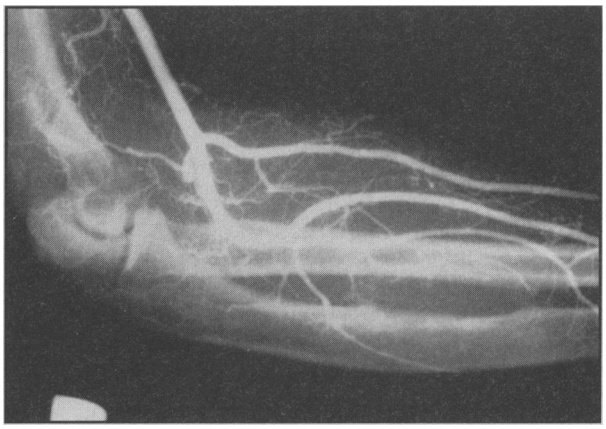

Figure 1B. Angiographic study of the right forearm and elbow demonstrating diffuse vascularity and contingent tumor involvement (Case 1).

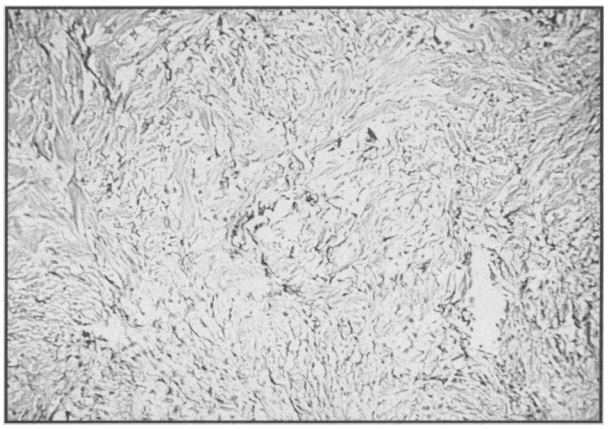

Figure 1C. The typical histologic pattern of aggressive fibromatosis demonstrating bundles of spindle cells associated with dense collagen in disarray (Case 1).

increased uptake in the soft tissue without apparent osseous involvement. Computerized tomography scans showed a lesion between the radius and ulna with a density similar to that of muscle or fibrous tissue and "erosion" of the ulnar cortex. By an angiograph, the early arterial phase revealed displacement of the proximal radial vessels by the tumor mass (Figure 1B). The mass itself remained relatively avascular. Other laboratory studies did not add significant information.

This patient underwent a wide excision[6] in November 1979, including resection of the radial half of the proximal portion of the ulna to which the tumor was densely adherent. The posterior interosseous nerve was intimately involved in the tumor mass, and it was cut at the proximal and distal margins of the resection and removed as part of the specimen. Histologically, the tumor consisted of bundles of spindle cells associated with large amounts of collagen (Figure 1C). There were few mitoses, minimal anaplasia, and no "herringbone" pattern typical of fibrosarcoma. The histologic diagnosis was aggressive fibromatosis.

Six months after surgery, the patient returned with persistence of the mass measuring 10×20 cm in the region of the previous surgery. It also had extended into the distal forearm over the ulna and radius. Radiographs showed "erosion" of the interosseous borders of both the radius and ulna. Arteriography demonstrated displacement of proximal forearm vessels but no neovascularity. The bone scan had increased activity throughout the forearm and carpus. A second procedure was accomplished in July 1980, but was marginal excision[6] and included resection of approximately 15 cm of the distal ulna and part of the medial cortex of the radius along with the interosseous membrane. Histologically, the tissue was identical to that of the original specimen.

Postoperatively, the patient received 5000 rads irradiation from the mid-metacarpal level to the elbow, crossing the joint. Two years after the initial procedure, the lesion appears to be controlled without clinical or radiological evidence of persistence. The patient has a functional upper extremity but uses a static wrist dorsiflexion brace with an outrigger to aid extensor action of the thumb and fingers.

Case 2

A 17-year-old white female was noted to have a deformity of her left upper extremity in early childhood.

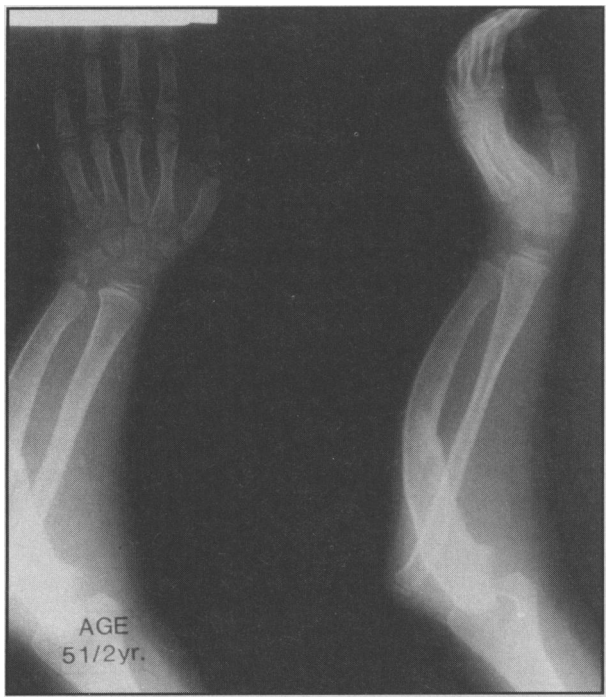

Figure 2A. Radiographs of the patient's elbow, forearm, wrist, and hand at age 5½ years. Distinct bowing of the ulna and congenital dislocation of the radius with middiaphyseal cortical irregularity are apparent (Case 2).

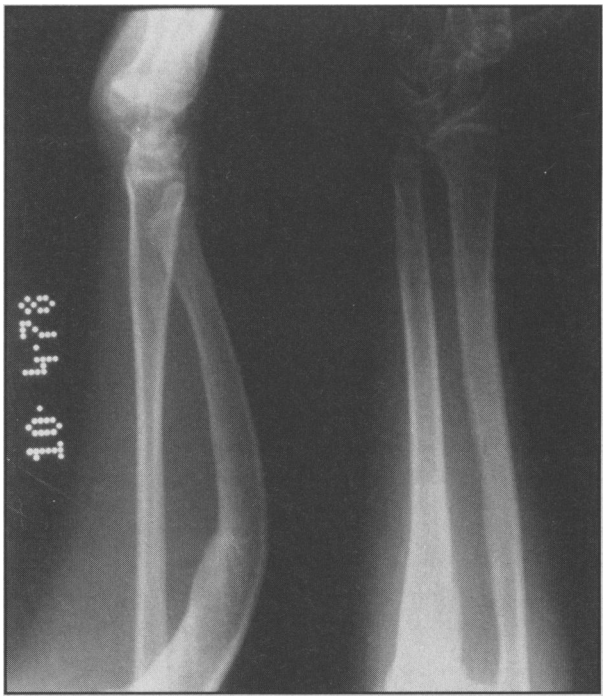

Figure 2B. Radiographs of the same patient's forearm and hand at skeletal maturity. Ulnar diaphyseal bowing persists. Early cortical changes are noted (Case 2).

Congenital dislocation of the radius with bowing of the ulna was radiographically documented at 5½ years of age (Figure 2A). In March 1979, she underwent an osteotomy and plating of the ulna and excision of the radial head for cosmesis and improved function (Figure 2B). The soft tissues at surgery were unremarkable.

After this procedure, the patient complained of persistent and increasing pain in the proximal extensor compartment. The pain consisted of two types: deep aching and a more superficial burning sensation throughout the lesion. The plate was removed in March 1980 because of the pain.

At surgery, a dense, fibrous tumor was found surrounding the ulna at the level of the osteotomy site. It had no distinct margins. An excisional biopsy was done, preserving major nerves and vessels. Pathologic diagnosis revealed aggressive fibromatosis.

After consultations with several authorities, the patient received 5000 rads of radiotherapy to the entire forearm in July 1980. Mild improvement lasted only several months (Figure 2C). The patient's pain returned to a level that required narcotic analgesia. Radiographs showed progressive destruction of both the radius and ulna. This destruction ultimately produced a pathological fracture of the ulna. In October 1981, the patient underwent a wide excision[6] in the form of an above elbow amputation. Grossly, the tumor had replaced the muscles and fascia of the proximal extensor compartment. Envelopment of the radial and ulnar nerves proximally and median nerve distally was pronounced. Histologically, the tissue was very similar to the previous specimen and was labeled aggressive fibromatosis (Figure 2D).

DISCUSSION

The development of an aggressive fibrous tissue lesion occurring secondarily to an ulnar deformity has not been reported previously, although an association of ulnar abnormalities and neurofibromatosis has been documented.[7] The occurrence of congenital fibrosarcoma in the forearm has been reported.[8] Hayry et al[9] in their extensive investigations, reported that 80% of patients with aggressive fibromatosis were found to have some minor skeletal anomaly upon radiographic screening. This led the authors to believe that a defect in connective tissue regulation could be an important underlying cause in the etiology of aggressive fibromatosis.[9]

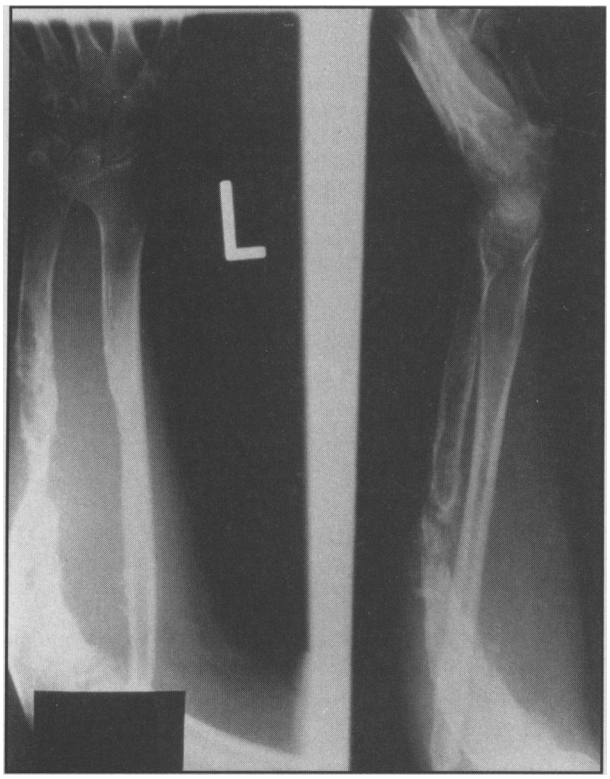

Figure 2C. Radiographs of the forearm and hand after radiotherapy. Destructive middiaphyseal resorptive changes are noted (Case 2).

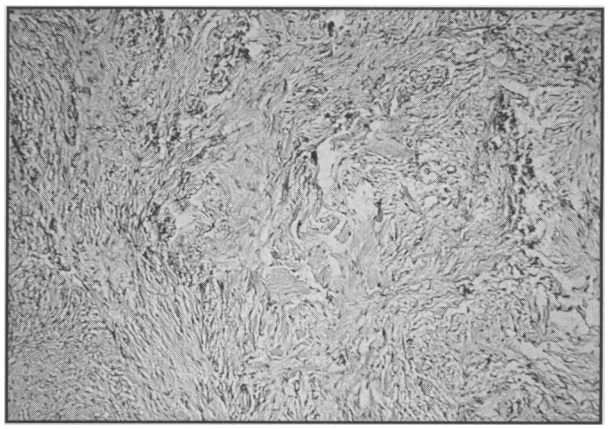

Figure 2D. Histological pattern of aggressive fibromatosis, characterized by a dense fibrous stroma, collagen in disarray, occasional cellular atypism, and intermediate vascularity is noted (Case 2).

The generic term "fibromatosis" was proposed by Stout[10] for a group of related conditions having the following features in common:

- proliferation of well-differentiated fibroblasts,
- infiltrative pattern of growth,
- presence of a variable amount of collagen between proliferating cells,
- lack of cytological features of malignancy and little mitotic activity, and
- aggressive clinical behavior characterized by local recurrences but without evidence of distant metastasis.

Numerous classifications of the fibromatosis have been proposed, with more recent literature acknowledging the clinical and pathological entity of aggressive fibromatosis. The term is frequently interchanged with extra-abdominal desmoids. Although the clinical and pathological features of the two are often indistinguishable, we prefer the term aggressive fibromatosis for these lesions because it reflects the true behavior and character of the lesion with greater precision.

From the original description of this general category of disease process by MacFarlane in 1932 to the present, the etiology of fibromatosis in general, and aggressive fibromatosis in particular, has remained obscure. Trauma, both surgical and nonsurgical, has been implicated with fibromatous lesions arising months to years later.[11,12] Endocrine abnormalities have found favor with some authors based on the relationship of abdominal desmoids to pregnancy[13] and the regression of an inoperable abdominal desmoid following irradiation castration.[10] It has been shown that the growth rate of aggressive fibromatosis directly correlates with the concentration of indigenous estrogen in females. Further, in a recent study correlating aggressive fibromatosis tumor growth rate with patient age and gender, Reitamo et al found that females ages 25 to 45 showed the most rapid tumor growth rate while premenarchal and postmenopausal females exhibited a growth rate similar to males of all ages.[14]

Historically, treatment of aggressive fibromatosis has been surgical, but the infiltrative nature of the lesion has resulted in inadequate excision and a significant persistence rate.[4,12,15-18] Moderate success in treating inoperable lesions with radiation has generated some enthusiasm for this mode of therapy for the primary lesion.[19] Preoperative chemotherapy followed by radical resection has been reported by Goepfert et al[20] for a mandibular lesion. The use of indomethacin and ascorbate in the treatment of desmoid tumors has recently been reported with limited follow-up.[21] Open biopsy followed by wide excision as a local procedure when the lesion is unicompartmental is our preferred treatment. Bicompartmental lesions or those involving critical neurovascular structures should be managed by

wide excision in the form of amputation. Persistence confined to a single compartment and away from critical neurovascular structures may render them amenable to excision short of amputation.

SUMMARY

These two case reports establish the association of aggressive fibromatosis with congenital bowing of the ulna. Both patients had pain as an early manifestation of their disease, which is unusual in most cases of aggressive fibromatosis. Both lesions were bicompartmental and had surrounded major nerves, explaining the nerve pain experienced by both patients. Both lesions aggressively involved the ulna as well as the radius to a lesser extent. The initial surgical procedures, while appropriate to preserve function in both cases, were inadequate to successfully control the lesion. Surgical intervention resulted in florid growth in both cases with no differences seen because of age, sex, or race. Radiation therapy has helped in controlling persistent tumor in one patient.

CONCLUSION

Aggressive fibromatosis may be a latent manifestation of congenital bowing of the ulna. The course of the disease appears to be an aggressive one, especially following surgical intervention. Any surgical procedure requires thorough preoperative planning, including identification of compartmental involvement to lessen the chance of persistence. Radiation therapy may have a role in selected cases. Close follow-up is imperative as not only recurrence but also radiation-induced malignant transformation of this lesion has been reported.

Acknowledgment

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